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Oral pemphigus.

[Article in English, Italian]

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Pemphigus is a group of potentially life-threatening autoimmune diseases characterized by cutaneous and/or mucosal blistering, due to the presence circulating IgG antibodies directed against desmoglein 1 and 3 (Dsg 1 and 3). Differences in the particular distribution of these result in different clinical manifestations of the disease. The most common variant is pemphigus vulgaris (PV). There is a fairly strong genetic background to pemphigus with linkage to HLA class II alleles and ethnic groups such as Ashkenazi Jews and those of Mediterranean and Indian origin are especially liable. Oral lesions are commonly characterized by the presence of vesiculobullous and ulcerative lesions. Diagnosis is achieved via three different parameters: perilesional tissue biopsy, histological and immunological examinations. Serum autoantibodies to either Dsg1 or Dsg3 are best detected using both normal human skin and monkey esophagus or by enzyme-linked immunosorbent assay. The main aim of treatment is to reduce inflammatory re-sponse and autoantibody production, in order to achieve disease remission in a short time. Before the advent of corticosteroids, PV was typically fatal due to dehydration or secondary systemic infections. Current treatment is largely based on systemic immunosuppression using corticosteroids, with azathioprine or other adjuvants or alternatives. Nonetheless, newer therapies, such as intravenous immunoglobulins (IVIg) or anti-CD20 monoclonal antibodies (Rituximab), with potentially fewer adverse effects also appear promising.

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